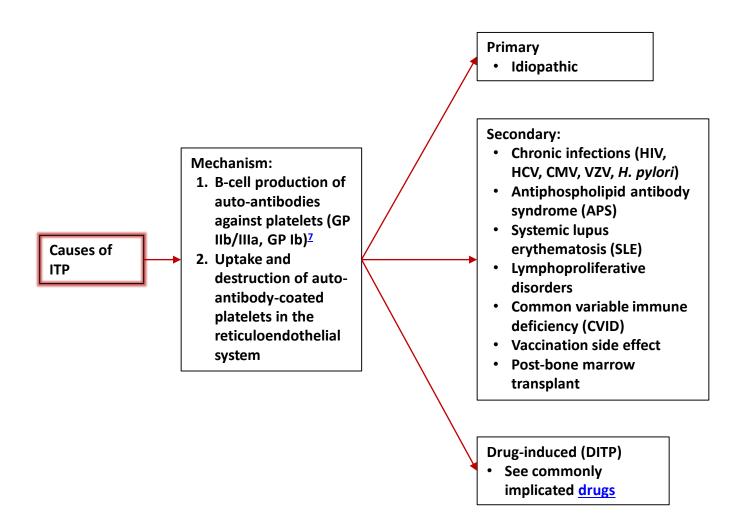
## Immune thrombocytopenic purpura (ITP)

**Updated: 5/24/2017** 

- 1. Causes of ITP and when to suspect
- 2. Workup for suspected cases
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### Causes of ITP and when to suspect



### Workup for suspected cases

**Essential history elements:** 

- Constitutional symptoms (fevers, chills, sweats, weight loss, lymphadenopathy)
- Signs of bleeding/bruising
- Alcohol use
- Intravenous drug use
- History of liver or renal disease
- Sexual history
- Recently-initiated drugs or supplements
- Diet

**Suspected ITP** 

 Family history of malignancy or bleeding disorders Minimum laboratory workup<sup>1,2</sup>:

- CBC with differential
- Peripheral smear
  - Platelet clumping?
  - Atypical cells (malignancy)?
  - Spherocytes (Evan's syndrome)?
  - Shistocytes (MAHA)?
- Coagulation tests (PT/INR, aPTT, fibrinogen)
- Liver tests
- Serum creatinine
- HIV serology
- HCV serology
- Ferritin, B12, folate

\*Bone marrow biopsy is NOT required to make the diagnosis of ITP<sup>1</sup>

Diagnostic criteria:

- Peripheral blood thrombocytopenia (<100 x 10<sup>9</sup>/L)
- Absence of other explanation for thrombocytopenia

\*ITP is a diagnosis of exclusion

Additional tests to consider on case by case basis:

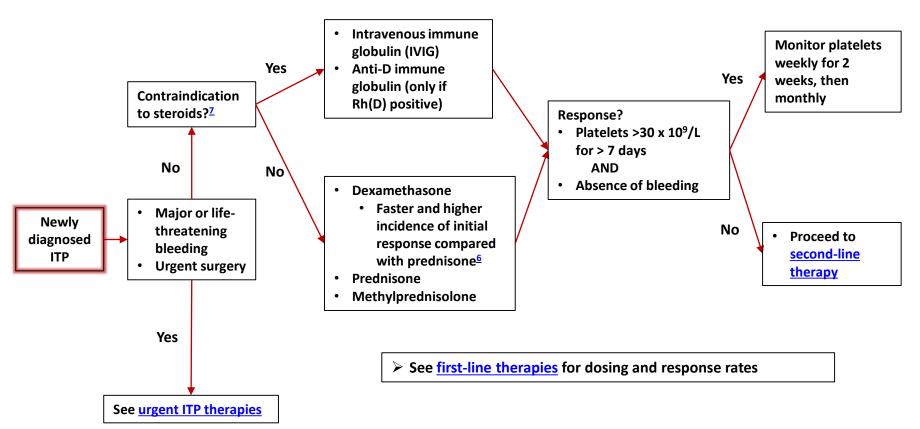
- Platelet refractoriness test (failure to increase platelets by >30,000 within 1 hour of transfusion, on two separate attempts)
- Stool H. pylori if GI symptoms
- ANA if suspected connective tissue disorder
- Reticulocyte count and direct antiglobulin (Coombs) if concurrent anemia
- Antiphospholipid antibodies if spontaneous arterial thrombosis, recurrent pregnancy loss
- Quantitative immunoglobulins if history of recurrent infections, allergies, or transfusion reactions
- Thyroid function tests if suggestive symptoms

Platelet auto-antibody assays:

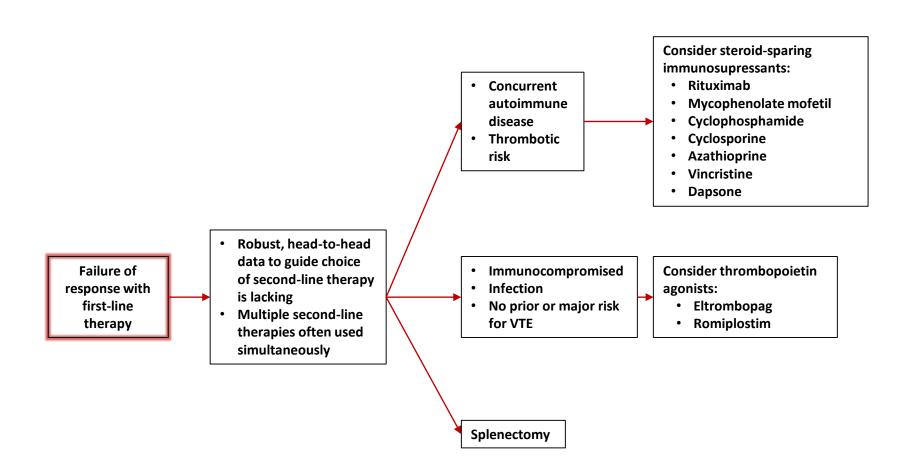
 Unhelpful due to poor sensitivity (40%)<sup>5</sup>

#### **Initial therapy**

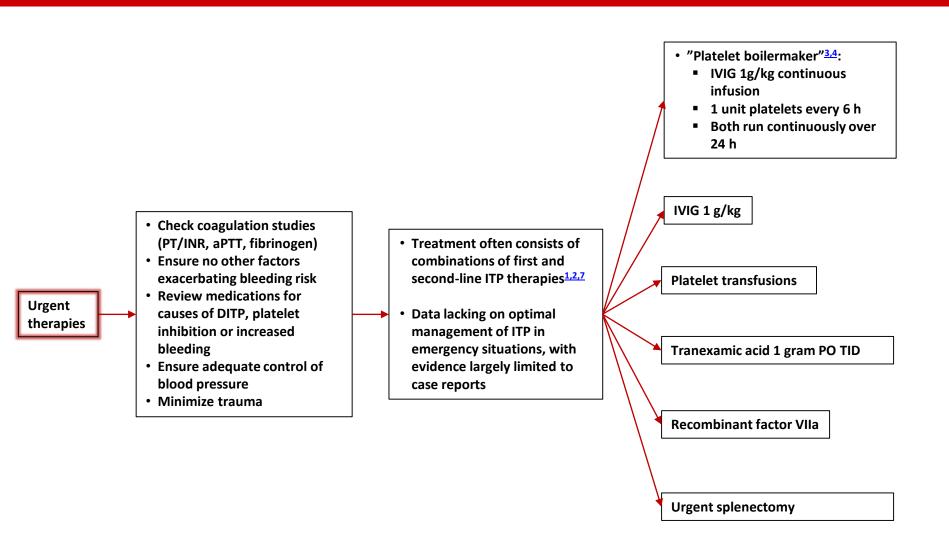
- General guidelines on who to treat<sup>1,2</sup>:
  - Platelets <30 x 10<sup>9</sup>/L OR
  - Bleeding signs or symptoms
- > Decision to treat must be individualized based on age, co-morbidities, bleeding risk, history of bleeding, activity level, and patient preferences.



## Second-line therapy 1.2.7



#### **Urgent therapies**



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# First-line therapies: dosing and efficacy<sup>1,2</sup>

Treatment	Initial response rate	Time to initial response	Peak response time	Duration of response	Toxicity
Corticosteroids  Dexamethasone  40 mg PO daily x 4 days, every 2-4 weeks for 1-4 cycles	Up to 90%	2-14 days	4-28 days	50-80% in remission after 2-5 years follow-up (using 3-6 cycles)	Weight gain, hyperglycemia, edema, hypertension, peptic ulcer disease, cataracts, avascular necrosis, immune suppression, adrenal
<ul> <li>Prednisone</li> <li>1 mg/kg/days x 40 days</li> <li>Methylprednisolone</li> <li>30mg/kg/d x 7 days</li> </ul>	70-80% Up to 95%	4-14 days 4-14 days	7-28 days	13-15% estimated remission at 10 years 23% in remission at 39 months	insufficiency
• 1g/kg/d for 1-2 days	Up to 80%	1-3 days	2-7 days	Short: platelets return to pre-treatment levels within 2-4 weeks	Flu-like illness, aseptic meningitis, neutropenia, thrombosis, rare anaphylaxis in IgA deficiency
Anti-D immune globulin • 50-75 μg/kg	Up to 80%	1-3 days	2-7 days	Short: platelets return to pre-treatment levels within 2-4 weeks	Hemolytic anemia, DIC, renal failure

# **Drug-induced ITP**

#### Commonly-implicated drugs

Class	Drug	
Heparins	UFH, LMWH	
Platelet inhibitors	Abciximab, tirofiban, eptifibatide	
Antibiotics	Linezolid, rifampin, vancomycin, sulfonamides	
Anticonvulsants	Phenytoin, carbamazepine, valproate,	
H <sub>2</sub> blockers	Ranitidine, cimetidine	
Analgesics	Acetaminophen, NSAIDS	
Diuretics	HCTZ, Chlorothiazide	

#### References

- 1. Neunert C, Lim W, Crowther M, et al. The American Society of Hematology 2011 evidence-based practice guideline for immune thrombocytopenia. Blood. 2011;117(16):4190-207.
- 2. <u>Provan D, Stasi R, Newland AC, et al. International consensus report on the investigation and management of primary immune thrombocytopenia</u>. Blood. 2010;115(2):168-86.
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- 4. Spahr JE, Rodgers GM. Treatment of immune-mediated thrombocytopenia purpura with concurrent intravenous immunoglobulin and platelet transfusion: a retrospective review of 40 patients. Am J Hematol. 2008;83(2):122-5.
- 5. Warner MN, Moore JC, Warkentin TE, Santos AV, Kelton JG. A prospective study of protein-specific assays used to investigate idiopathic thrombocytopenic purpura. Br J Haematol. 1999;104(3):442-7.
- 6. Wei Y, Ji XB, Wang YW, et al. High-dose dexamethasone vs prednisone for treatment of adult immune thrombocytopenia: a prospective multicenter randomized trial. Blood. 2016;127(3):296-302.
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